Spinal Dysraphism: A Neurosurgical Review for the Urologist

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Spinal neural tube defects are congenital malformations of the spine and spinal cord (eg, myelomeningocele) and are frequently seen in pediatric urology practice. These neurologic problems have many consequences in a child's life and affect different parts of the body, such as the brain, spinal cord, limbs, bladder, and bowels. Because of the complexity and neurologic aspects of spinal dysraphism, many related terms and aspects of the disease are unfamiliar to the urologist. This review addresses some of the most commonly used neurosurgical terms and concepts related to spinal dysraphism.
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Spinal neural tube defects are congenital malformations of the spine and spinal cord secondary to abnormal neural tube closure that occur between the third and fourth weeks of gestation. The term *spinal dysraphism* includes the overall group of defects derived from the maldevelopment of the ectodermal, mesodermal, and neuroectodermal tissues, and its sequelae may affect brain, bones, extremities, and bowel and bladder functions.

The incidence of spinal dysraphism ranges from 3.2 to 4.6 per 10,000 births in North America[1,2]; no geographic variation has been seen, and there is a relatively

uniform incidence in all ethnic groups.[3] There is strong evidence that there has been a decline in incidence worldwide since the 1970s[1,4]; however, it is unclear whether this is a transient or permanent trend. This decline is probably due to a systematic use of dietary folic acid before and during the gestational months,[5] and more recently to the advent of prenatal diagnosis, which leads to therapeutic abortion in as many as half of the diagnosed cases in some countries.[6] The disorder occurs equally or somewhat more commonly in female newborns (female, 1.0-1.7/male, 1.0), depending on the populations studied. Embryologically, open spinal dysraphism (myelomeningocele) is thought to occur 3 to 4 weeks after conception at the time that the neural tube is closing.[7]

Myelomeningoceles are by far the most common spinal dysraphic condition affecting the lower urinary tract and therefore the most familiar to urologists.[8] The lumbar and sacral regions are the most common vertebral levels affected[9] (Table 1).

Before advances in ventricular shunting devices, the survival of a child with open spinal dysraphism was dismal, and, therefore, urologic intervention was rarely necessary. As the life expectancy of these children increased, so did the morbidity and mortality secondary to urologic complications, such as pyelonephritis, hydronephrosis, and renal failure.[10,11] The need for appropriate urologic evaluation and effective management became mandatory to improve the health, longevity, and quality of life of patients.

The evaluation and management of the neurogenic lower urinary tract secondary to spinal dysraphism has undergone a major evolution over the past 30 years.[12-19] This has been fueled by advances in urodynamic technology and an improved understanding of the long-term effects of a urodynamically hostile bladder and bladder outlet.[20-22] At the same time, improved methods for maintaining a low-pressure bladder reservoir and providing for adequate emptying through intermittent catheterization has resulted in a marked improvement in prognosis.[23-34]

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In addition to the urologic problems, patients with spinal dysraphism often have other systemic disorders that require medical attention by a multidisciplinary approach. We focus this article on the premise that a sound understanding of the neurologic aspects of spinal dysraphism will lead to enhanced outcomes for affected children. We discuss neurologic terminology, epidemiology, etiologic risk factors, associated congenital anomalies, prognosis, and appropriate neurosurgical evaluation and management. The urologic evaluation and management of the spinal dysraphic bladder will not be discussed and has previously been extensively reviewed.

Table 1
Vertebral Levels of Myelomeningoceles

Level	Estimated Prevalence (%)
Cervical	0-5
Thoracic	5-10
Thoracolumbar	20-30
Lumbar	20-30
Lumbosacral	30-50
Sacral	5-15

Data from Bauer SB et al.[9]

Neurologic Terminology

The term *spinal dysraphism* is general and represents an expansive list of neurologic disease entities that may be unfamiliar to the urologist; we tend to group all spinal defects together, inappropriately referring to them as *myelodysplasia* or *myelomeningocele*. The term *spinal dysraphism* is more appropriate when describing children with a vast array of congenital spinal abnormalities. Specific terms are defined in Table 2, and some are illustrated in Figure 1. Because myelodysplasia has a limited focus that includes closed defects of the spinal cord or roots, and more importantly is a term often used to describe a multitude of hematologic dyscrasias, we believe that myelodysplasia should not be used at all to describe patients with spinal dysraphism.

Many patients with spinal dysraphism have more than a single neurologic condition. When possible, one should describe these conditions specifically (eg, "an 8-year-old ambulatory girl, born with L2-L4 myelomeningocele, hydrocephalus shunted since birth, a stable Chiari II malformation, and a symptomatic tethered cord released a year ago"). In this manner, one may better understand the child's neurosurgical condition at a particular point in time.

Myelomeningocele

Myelomeningocele is the most common dysraphic malformation and occurs in approximately 1 in 1200 to 1400 births.[35] Myelomeningocele derives from a failure of the neural tube to close or a secondary reopening of the closed neural tube.[36]

Table 2
Common Neurologic Terminologies of Spinal Dysraphism

Term	Definition
Arthrogryposis	Congenitally absent anterior horn cells with resultant weakness and contractures.
Chiari II malformation	Hindbrain and cortical malformation seen almost exclusively with myelomeningocele patients (noted in 50%-90% of patients; less frequent in those with lower-level lesions). Only severe cases (approximately 10%) affect the cardiopulmonary system and/or cranial nerves and upper extremities, which require emergent neurosurgical intervention. The hindbrain hernia consists of caudal migration of the cerebellar vermis and lower brainstem. The cortical malformation has many components involving the cortex, basal ganglion, and upper brainstem. Examples of involvement include enlargement of the massa intermedia of the third ventricle, beaking of the tectum, polygyria, hypoplasia of the tentorium, and flax cerebri.
Chiari III malformation	Posterior fossa encephalocele, which contains cerebellar and brainstem tissue. This is the most severe form of hindbrain herniation and is often lethal.
Split cord anomalies (diastematomyelia)	Splitting of the spinal cord with or without a bony or cartilaginous septum. Below the level of the lesion the legs are usually involved asymmetrically. Frequently the level of the lesion is marked by focal hirsutism. This is such a predictable finding that if no split cord is found the study should be scrutinized again. Differs from diplomyelia, in which true duplication of the cord into "twin cords" occurs without bony or cartilaginous spurs.
Encephalocele	Herniation of the brain and meninges through a defect in the skull (75% in the occipital region); it is a cranial meningocele only if the meninges are involved (with a more benign prognosis).
Hydrocephalus	Enlargement of the brain's ventricular system (found in approximately 90% of patients with thoracolumbar and lumbar level lesions, 75% of lumbosacral lesions, and 50% of sacral lesions). Related to the Chiari II malformation (communicating) or the Sylvian aqueduct stenosis (noncommunicating).
Lipomyelomeningocele	Fatty infiltration of the distal spinal cord, usually associated with a subcutaneous lipoma. The fat may enter the dorsal or caudal aspect of the cord and may or may not have a thickened filum seen with it. Urologic problems are frequently the initial presentations of the patient. The natural history of this lesion is one of progressive loss of neurologic function. It is the most common type of intraspinal tumor in the spinal dysraphism population (other types include chondromas, osteomas, angiomas, and dermoids).
Lumbosacral agenesis	Rare. Complete or partial absence of lumbar spine and sacrum; paraplegia and orthopedic lower limb deformities are common.
Meningocele	A sac containing cerebral spinal fluid and no neural elements. This is extremely rare, being present in $<$1% of all neural tube defects of the spine. When seen, the clinical course is usually quite benign, without neurologic abnormality.
Meningocele manqué	A meningocele that failed to develop fully. This term usually refers to strands of neural tissue coming from the medial aspect of a split cord anomaly fixating the cord dorsally.
Myelomeningocele	Spina bifida cystica with neural elements (most common etiology of pediatric neuropathic bladder). Although the term *meningomyelocele* is synonymous with myelomeningocele, neurosurgeons by convention prefer the latter term.
Rachischisis	A cystic degeneration of vertebral bodies and cord.
Spina bifida aperta	Spina bifida with an open cord on the skin surface, without cystic covering. Sometimes covered by thin membranes. Synonymous with myeloschisis, myelocystocele, and myelocele.
Spina bifida cystica	Spina bifida with a cystic lesion on the back consisting of dura, meninges, and normal or abnormal skin.
Spina bifida manifesta	Spina bifida with skin surface manifestations, such as hemangioma, hair, sinus tract, and covered or open neural elements.
Spina bifida occulta	Spina bifida (usually L5-S1) with normal meninges and neural elements (found in 5%-10% of neurologically normal adults and in up to 50% of newborns before ossification). Includes meningocele manqué, lipomeningocele, diastematomyelia, and dermal sinus or cyst (extra- or intramedullary fistulous tract or cyst—a potential source of meningitis).
Spinal dysraphism	Incomplete closure of the neural tube (almost a global term).
Syringomyelia	Cystic degeneration of the cord.

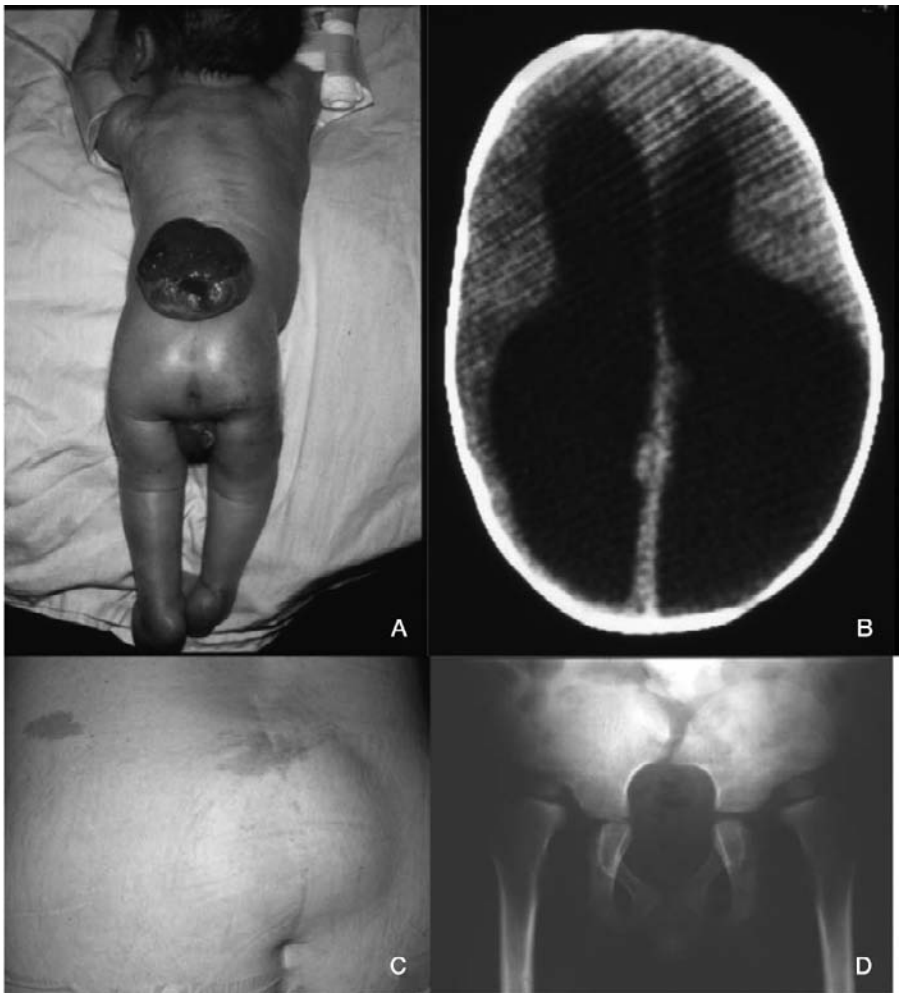

Figure 1. (A) *Lumbar myelomeningocele;* (B) *computed tomography showing a hydrocephalus;* (C) *lipomyelomeningocele (sacral lipoma); and* (D) *radiograph demonstrating a sacral agenesis.*

The term *myelomeningocele* is used to describe open spinal dysraphism. There is no such thing as closed myelomeningocele. It can occur at any level of the spinal cord and is the most severe form of dysraphism.

The majority of children (80%) with isolated myelomeningocele have normal intelligence, although 60% have some learning disability. The life expectancy of these children is nearly normal.[37] Most of these children (60%) are community ambulators, and 80% are socially continent (dry), although many of them receive clean intermittent catheterization.[38]

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Myelomeningocele is a static disease; any deterioration in these children must be examined carefully, and a clinical evaluation and imaging study should be done promptly. The most common cause of deterioration is shunt malfunction. Other causes are tethered cord, Chiari malformation, and syringomyelia. The most frequent symptoms of deterioration are headache, nausea, vomiting, behavior modification, and change in upper or lower extremity strength and coordination. The urologist must be aware and pay close attention to modifications in urinary function and bowel habits. Frequently a change in bladder function detected in routine urodynamic study may lead to diagnosis of a tethered cord.

Occult Spinal Dysraphism

Occult spinal dysraphias are closed forms of spinal dysraphism in which the skin covers the neural tissue. They occur most often at S1, S2, or both.[39] Although some of these spinal dysraphic lesions are truly occult, in most a skin marker is present (hairy patch, cutaneous nevus, an appendage or skin tag, small dimple with a pinhole, lipoma).[40] Recognizing these cutaneous marks is important because they are usually associated with some form of dysraphism that can cause spinal cord injury and lead to progressive and sometimes sudden neurologic deterioration (Table 3). Stabilization of the lesion may be achieved by untethering the cord, but neurologic, urologic, and orthopedic problems are often irreversible when they occur.[37] Therefore, most pediatric neurosurgeons prefer to correct these malformations prophylactically, before the onset of symptoms.

Occult spinal dysraphias may be of many different embryologic etiologies, but they are usually associated with tethering of the spinal cord.[41,42] The most common occult forms of spinal dysraphism are lipomas, split cord malformations (diastematomyelia and diplomyelia), dermal sinuses and dermoid tumors, myelocystoceles, tight filum terminale, neuroenteric cysts, and caudal agenesis.

Table 3
Skin Stigmata of Occult Spinal Dysraphism

Skin Stigmata	Associated Condition
Sacral dimple	Dermal sinus tract
Hairy patch	Diastematomyelia
Nevus	*
Capillary hemangioma	*
Central sacral mass of lipoma	Lipomyelomeningocele
Dermal sinus tract above gluteal crease	*
Absent or asymmetric gluteal cleft	Sacral agenesis
Skin tag or tail-like appendage	*
Atretic myelomeningocele scar ("cigarette burn")	*

*Nonspecific occult tethered cord.
Data from Dias MS and Li V[37] and Drolet BA.[109]

Hydrocephalus

Hydrocephalus is not a specific disease; rather, it represents a diverse group of conditions. In the case of myelomeningocele it results from impaired circulation and absorption of cerebral spinal fluid. Myelomeningocele and cerebral hemorrhage of prematurity are the leading causes.[43,44] Hydrocephalus occurs in approximately 85% of children with myelomeningocele and bears little relationship to intelligence.[45,46] Generally the chance of developing hydrocephalus is greater in upper lesions. It is uncommon in the closed forms of spinal dysraphism.

Clinical manifestations of hydrocephalus vary with age and include enlargement of the head, vomiting, irritability, and lethargy. Headache may be present in older children. An ultrasound or a computed tomography scan or magnetic resonance imaging (MRI) can confirm the diagnosis.

Hydrocephalus does not directly affect the urologic course of the patient, other than when major intra-abdominal procedures are performed in the presence of an indwelling ventriculoperitoneal shunt. The risk of infecting the shunt should be considered in these cases.

Chiari II Malformation

In Chiari II malformation the posterior fossa is small, and the cerebellum, pons, and medulla are displaced to varying degrees into the cervical canal, leading to a variable degree of compression of the brainstem, which may be caused by an abnormal development of the ventricular system because of the open dysraphism.[47] Some element of Chiari II malformation is present in most children with a myelomeningocele.

Respiratory and swallowing difficulties associated with Chiari II are the primary causes of morbidity and mortality in the first 2 decades of life in this group of patients.[48] Shunt dysfunction or untreated hydrocephalus can mimic all the symptoms of hindbrain compression. Differential diagnosis has to be made before starting management. The treatment for Chiari II malformations is surgical decompression of the hindbrain in the cervical canal.

Patients with occult spinal dysraphism or the closed form of neural tube defects rarely have the changes of the Chiari II malformation, and less than 3% will have caudal descent of the cerebellar tonsils (Chiari I) without any change in the cortical architecture.

Tethered Spinal Cord

During normal fetal development the bony spine grows at a greater rate than the spinal cord. This difference in rate results in a progressive disparity between the termination of the spinal cord and that of the bony spine. At 8 weeks' gestation the conus medullaris ends at the coccygeal vertebral level. By 24 weeks' gestation it lies at the L3-L4 level. Not until approximately 2 months after birth does the conus medullaris come to lie at the permanent adult vertebral level of L1-L2.[49-52]

The spinal cord fixation commonly referred to as "tethered cord" may be a result of a variety of conditions, including adhesions from a previously repaired myelomeningocele, lipoma of the caudal spinal cord, and split cord anomalies such as diastematomyelia, meningocele manqué, and other conditions. The clinical manifestations of spinal cord fixation syndromes are believed to result from an ischemic event, usually caused by stretching of the spinal cord, with early surgical release allowing the best chance for neurologic recovery.[53]

The incidence of retethering in the myelomeningocele population has been estimated at 15% to 20%.[54] Its diagnosis is primarily clinical, with patients presenting with progressive or subtle loss of function, and it is usually detected by careful and regular evaluations.

It is important for urologists to recognize the presence of a tethered cord because it may present as new-onset or a pattern change of voiding dysfunction in this population. Numerous reports have shown urodynamic improvements in some patients after

Table 4
Conditions Requiring Screening for Spinal Dysraphism

Condition	Incidence (%)
Anorectal malformation	17-34
VATER syndrome	10
Cloacal exstrophy	100

Data from Appignani BA et al[110] and Botto LD et al.[111]

surgical release of the fixed spinal cord.[55-62]

Screening for a tethered cord. Patients at risk for a tethered cord include those with cloacal exstrophy, imperforate anus, VATER syndrome, and cutaneous stigmata of occult dysraphism (focal hirsutism, midline dermal sinus above the gluteal crease, subcutaneous lipoma, capillary hemangioma, midline appendages, dermal dysplasia resembling a "cigarette burn"), among others (Tables 3 and 4). It is recognized that up to 10% to 50% of patients with surgically significant occult spinal dysraphism will have normal skin; therefore, screening for intradural pathology only on the basis of skin inspection is a poor method of detection.[63]

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The majority of myelomeningocele patients have radiographic evidence of a tethered cord on MRI. Therefore, radiographic evidence alone is not a justification for operation. Patients with symptoms referable to the area, particularly if the problems are progressive, should be considered candidates for operative detethering. Symptoms may be subtle and may simply be a change in the continence pattern or a worsening in scoliosis.

Children with voiding dysfunction are a mainstay of urologic practice. Evaluation of all of them by MRI looking for a neurologic cause is inappropriate and costly. There are some criteria that will enhance the yield. Any patient with cutaneous stigmata of occult dysraphism should be imaged, whether symptomatic or not. This implies that the skin of the back should be examined. Any child with neurologic deficit or back or leg pain should also be imaged. Those with a neurogenic pattern to their urodynamic study or significant bony dysmorphism should be considered.

Appropriate imaging of the intradural anatomy can be accomplished in a child up to 4 to 6 months of age by ultrasonography.[64,65] Premature children should not be screened until they reach full-term gestational age because of the naturally low position of the conus. After 6 months of age, MRI is the most appropriate imaging study. Computerized tomography with or without contrast is reserved for difficult, confusing anatomy screened by MRI or where MRI is not available. Conventional radiographs of the lumbar spine may add additional information concerning the segmentation and dorsal bony anatomy of the spine, but cannot be used to screen patients for surgically significant pathology.

Etiologic Risk Factors

Although no clear etiology is known to result in either the open or closed forms of spinal dysraphism, some regional adverse factors have been reported, primarily involving the mother at conception and early pregnancy. Approximately 50% of cases are related to nutritional deficiency[66]; the remaining cases, which are inherited, are multifactorial. Some of the other causes are chromosomal abnormalities, single-gene abnormalities, environmental factors,[67] or are unknown. The ingestion of cytochalasin, a metabolite of the fungus *Phytophthora infestans* (found in blighted potatoes), folic acid or zinc deficiency, high nitrates (eg, nitrate-cured meats, bore and ground water), and vitamin A deficiency or excess have all been shown to be possible maternal nutritional causal elements.[68,69] An altered carbohydrate metabolism (eg, diabetes mellitus, hyperinsulinemia, or insulin-albumin antagonism) has been reported to be present in mothers of children with spinal dysraphism, particularly those with sacral agenesis. Mothers with diabetes are more prone to give birth to children with spinal dysraphism.[70]

One of the most important nutritional factors related to the advent of spinal dysraphism is the lack of folic acid. The use of supplementary folic acid may reduce neural tube defects by up to 72%.[68]

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Although no association with socioeconomic status has been well documented, significant evidence exists to support the importance of genetic factors in the development of spinal dysraphism.[71] There is a 3-fold higher incidence in consanguineous marriages, as well as a higher incidence in monozygotic twins. The mother of an affected child is 50 times more likely to have a second affected child (ie, a 5% chance) and

is 100 times more likely (ie, a 10% chance) if she has had 2 previously affected children.[2]

Recommendations During Pregnancy

During pregnancy, mothers are advised to avoid hyperthermia (eg, fevers, hot saunas or baths), as well as several medications (eg, valproic acid, clomiphene, and folic acid antagonists, such as aminopterin). The dietary folic acid supplement is recommended to be 0.4 mg daily for all women of childbearing age and is 10 times that amount (4.0 mg daily) if there has been a previous pregnancy with an affected fetus.[72]

Associated Congenital Anomalies

Table 5 depicts the most common urologic anomalies present in patients with spinal dysraphism. Orthopedic, craniofacial, cardiopulmonary, and gastrointestinal anomalies are also common (eg, congenital hip dislocations, equinovarus or paralytic clubfeet, kyphosis, scoliosis, atrial or ventricular septal defects, pilonidal cysts, imperforate anus, fecal incontinence and constipation). A lesion tethering the spinal cord is found in more than 50% of patients with anorectal, urogenital, or sacral malformations.[73]

Prognostic Factors

The overall medical and psychosocial prognosis of patients with spinal dysraphism depends on the extent of the neurologic deficits and associated congenital abnormalities, as well as the extent and sophistication of the treatment they receive. In general, the lower and less severe the spinal lesion, the higher the chance the patient will be ambulatory and not have hydrocephalus and, therefore, a better outcome.

Children with spinal dysraphism seem to have a higher risk for exhibiting worse levels of internalizing symptoms and lower levels of self-esteem than normal children.[74,75] They are also more likely to be interpersonally lonely and socially immature.[76] Spinal dysraphic children, especially those with hydrocephalus, frequently have difficulties in certain academic areas, such as arithmetic,[77,78] and they tend to score at the low end of the average range of intelligence. They also tend to exhibit deficits in executive functioning, abstract reasoning, and the ability to focus attention.[79] Parents who have a positive and hopeful attitude are able to improve the quality of life of their adolescents by up to 25% over that which would be predicted for the disability at birth.[80]

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The status of the lower and upper urinary tracts primarily depends on the individual patient's neurologic condition.[81] At birth, it is believed that 5% to 25% of children with spinal dysraphism will demonstrate an abnormal upper urinary tract (mostly mild reflux),[82] with up to 3% having decreased renal function (significant hydronephrosis). In a series of 64 infants, 9 patients (14%) were born with abnormal upper urinary tracts, with an additional 6 (9%) subsequently developing upper tract deterioration within 3 years of life.[83] If untreated, 10% to 50% of patients will develop not only abnormal upper tracts but also significantly decreased renal function. Therefore, appropriate management of these individuals may prevent significant urologic morbidity and mortality from taking place.

The life expectancy of patients with significant neurologic lesions is shorter than that of the general population. It is estimated that approximately 40% to 50% of children with neural tube defects will die during infancy.[81,84] In the past, prolonged life expectancy was almost exclusively achieved by ambulatory patients with sacral lesions and without hydrocephalus. If patients survived their

Table 5
Urologic Anomalies in Spinal Dysraphism[82,112-121]

Disorder	Prevalence in Spinal Dysraphism (%)	Prevalence in General Population (%)
Vesicoureteral reflux	21-25	2-31
Cryptorchidism	10-30	1.8-4
Bladder exstrophy	6.8	0.03
Hypospadias		0.2-0.4
Unilateral renal agenesis	2-8	0.05-0.1
Ureteropelvic junction obstruction	1-3	0.07-0.1
Multicystic dysplastic kidney	7-13	0.2
Horseshoe kidney	2-7	0.3

neurologic problems, life span depended mostly on their subsequent renal function. Therapy for hydrocephalus and antibiotics, developed in the 1950s, had the most significant impact on patient survival, because hydrocephalus was the major source of infant mortality. At all ages, renal failure is the most common cause of death. In children, the risk of renal failure is strongly related to the sensory level.[85] Renal failure is rare with sensory levels at or below L4, and common at or above T10.[86]

Adulthood brings little, if any, relief from the burdens of the patients with myelomeningocele. Anything that is not resolved in childhood will be more difficult to manage in adulthood.[85] As the child grows there is a natural tendency for the physical anomalies to deteriorate. The spinal deformity becomes more pronounced. Those who could just walk tend to relapse into a wheelchair.[87] Patients with spinal dysraphism are more prone to present with atherosclerosis, even if not obese.[88]

Once the child reaches young adulthood, concerns regarding sexual function and fertility begin. The physical aspects of sexuality that depend on the brain are usually intact, whereas those that rely on the spinal cord are compromised. In general, a complete or significant spinal cord lesion results in genital anesthesia. Male patients with significant sacral lesions (eg, no bulbocavernosus or anocutaneous reflex) are at higher risk for erectile dysfunction. Reflex erections are possible if the lesion is above the sacral region. All patients with intact sacral reflexes and urinary continence are potent.[85] Patients with suprasacral lesions are at a somewhat higher risk of ejaculatory or organic dysfunction with ejaculation and orgasm.[89] Incomplete or minor lesions are less likely to result in male sexual dysfunction. The management of sexual dysfunction is the same as in the normal population. Male fertility depends on erectile ejaculatory ability as well as the history of cryptorchidism. A great number of spinal dysraphic men are azoospermic.[90] Most women with lesions below L2 are thought to have normal sexual sensation, and 20% with higher levels have normal sexual function.[91] Fertility is not generally affected in women, but pregnancy is usually difficult and with higher risk of spinal dysraphism in the offspring.[92] The sexual libido and function of women with spinal dysraphism has not been as well documented as that of men. Endocrine function in both sexes is usually normal. There have been reports of spinal dysraphic children having an increased incidence of precocious puberty.[93,94]

The long-term survivorship (> 25 years) has anecdotally improved, although without reasonable statistics for patients with suprasacral lesions. Although the social and economic impact of improved life expectancy is not well documented, approximately 75% of adult survivors may be dependent on parents or other providers.

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Latex Allergy

Descriptions of apparent allergic reactions to natural rubber appeared in the medical literature in 1927, and irritant and delayed-contact reactions were reported in 1933.[95] The first report of latex allergy in a patient with spinal dysraphism was published in 1989,[96] and from then on an increasing number of cases have been recognized and published. The prevalence of latex allergy in patients with spinal dysraphism is high and ranges from 20% to 40%.[97-99] Latex allergy is an immunoglobulin E–mediated hypersensitivity reaction, and its symptoms range from mild urticaria to life-threatening events (bronchospasm, laryngeal edema, and systemic anaphylaxis) and death.

Latex antigen exposure can occur by cutaneous, percutaneous, mucosal, and parenteral routes, and the antigen can be transferred by direct contact and aerosol, but it is clear that direct mucosal and parenteral exposure poses the greatest risk of anaphylaxis.[100] It has been suggested that the most important factor in latex sensitization is the degree of exposure.[101-102] The number of surgical procedures and exposure episodes were the dominant factors in the development of latex allergy among children with spinal dysraphism, particularly as neonates and infants.[98,103] Conversely, there seemed to be no increased risk of latex allergy associated with age or sex.[104]

Children with other diseases requiring multiple surgical exposures with latex materials seem less prone to sensitization than children with spinal dysraphism,[105-107] and it has been suggested that there is a genetic association between spinal dysraphism and latex sensitivity.[106,108] Therefore, it is our belief that all children with spinal dysraphism, especially those undergoing multiple exposures to latex, should avoid subsequent contact to latex whether in the home, office, or hospital environment.

The operative risk of severe reactions is not as high in those patients without a history of latex sensitivity. Patients with a history of latex reactions can be safely treated with avoidance of equipment containing latex

and premedication. A careful history of latex sensitivity should be investigated in all patients with spinal dysraphism and, for those with latex allergy, appropriate safeguards should be maintained during their hospitalization by avoiding latex-containing equipment, gloves, and catheters. ■

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Main Points

- Myelomeningocele is the most common dysraphic malformation and occurs in approximately 1 in 1200 to 1400 births. Most of those children (60%) are community ambulators, and 80% are socially continent.
- Occult spinal dysraphias are closed forms of spinal dysraphism in which the skin covers the neural tissue. In most cases a skin marker is present. Recognizing these cutaneous marks is important because they are usually associated with some form of dysraphism that can cause spinal cord injury and lead to progressive and sometimes sudden neurologic deterioration.
- Hydrocephalus occurs in approximately 85% of children with myelomeningocele; it does not directly affect the urologic course of the patient, other than when major intra-abdominal procedures are performed in the presence of an indwelling ventriculoperitoneal shunt.
- The spinal cord fixation commonly referred to as *tethered cord* may be a result of a variety of conditions. It is important for urologists to recognize the presence of a tethered cord because it may present as new-onset or a pattern change of voiding dysfunction in this population. One of the most important nutritional factors related to the advent of spinal dysraphism is the lack of folic acid. The use of a supplementary folic acid may reduce neural tube defects by up to 72%.
- At birth, it is believed that 5% to 25% of children with spinal dysraphism will demonstrate an abnormal upper urinary tract (mostly mild reflux), with up to 3% having decreased renal function (significant hydronephrosis).
- In general, a complete or significant spinal cord lesion results in genital anesthesia. Male patients with significant sacral lesions (eg, no bulbocavernosus or anocutaneous reflex) are at higher risk for erectile dysfunction. Patients with suprasacral lesions are at a somewhat higher risk of ejaculatory or organic dysfunction with ejaculation and orgasm.
- Most women with lesions below L2 are thought to have normal sexual sensation, and 20% with higher levels have normal sexual function. Fertility is not generally affected in women, but pregnancy is usually difficult and with higher risk of spinal dysraphism in the offspring.
- The prevalence of latex allergy in patients with spinal dysraphism is high and ranges from 20% to 40%. All children with spinal dysraphism, especially those undergoing multiple exposures to latex, should avoid subsequent contact to latex whether in the home, office, or hospital environment.

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